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Leiomyosarcoma of the Mesenteric Root: A Strategic Location of a Rare Tumor

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Key Words
Leyomiosarcoma · Mesentery

Abstract
High mesenteric root sarcomas are difficult to manage due to their proximity to the superior mesenteric vessels. Resection of these tumors along with the blood vessels may lead to a complicated and protracted convalescence for the patient. Resection remains the main treatment modality for these tumors. During operation on high mesenteric root sarcomas, sound clinical judgment is needed for the decision not to sacrifice vital blood vessels.

Introduction

Retroperitoneal sarcomas are relatively uncommon tumors, constituting only 10–15% of all soft tissue sarcomas (STS) [1, 2]. The average annual incidence of these sarcomas is approximately 2.7 cases per million [3].

Patients usually present in their fifth decade of life, although the age range is wide. Males and females are equally affected [4–6].

The most common histological types of retroperitoneal STS are liposarcomas and leiomyosarcomas followed by pleomorphic undifferentiated sarcoma and malignant fibrous histiocytoma. A variety of other histological types exist but they are much less common in the retroperitoneum than in other primary sites.

Primary retroperitoneal or intraperitoneal leiomyosarcomas are very rare tumors. Here, we describe a patient who presented with a large mesenteric root leiomyosarcoma that was treated surgically with favorable outcome.

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Case Presentation

A 65-year-old male presented to our department complaining of recurrent epigastric and upper right abdominal pain radiating to the back for the last 3 months. No other complaints were prominent. The patient had undergone an uneventful laparoscopic cholecystectomy 2 years earlier. His family history was consistent with a father who suffered lung malignancy and a mother who expired due to hepatoma.

An ultrasound performed prior to his admission revealed a hypoechoic 4 × 4 cm lesion close to the pancreatic head. On admission, abdominal examination revealed a soft, nondistended abdomen with local tenderness in the upper right quadrant. No other findings were noted. His blood tests were all within normal limits. CEA and CA 19-9 were normal. Abdominal tomography revealed a 5 × 5 cm, rounded, soft density lesion adjacent to the root of the superior mesenteric artery (SMA) and pancreatic head (fig. 1). It was not possible to distinguish the origin of this lesion.

The patient underwent endoscopic ultrasound examination that demonstrated a hypoechoic-heterogeneous solid lesion behind and adjacent to the SMA and superior mesenteric vein. No other pathology was demonstrated in the pancreas itself (fig. 2).

Pathology obtained by fine needle aspiration from the lesion revealed fragments of mesenchymal tissue that was positively stained for c-kit and actin. DOG immunohistochemistry was negative. Laparotomy revealed a large, 7-cm, hard tumor adherent to the medial border of the pancreatic uncinate process laterally, to SMA medially, posterior to the portal vein and anterior to the inferior vena cava and left renal vein (fig. 2). The lesion was successfully dissected from these structures and was completely excised and removed.

The patient was discharged after an uneventful postoperative course. The pathological report revealed leiomyosarcoma stained to actin and desmin. The proliferative index Ki67 was 10%. He received radiotherapy, and at 4 years of follow-up, the patient is healthy and has no recurrence.

Discussion

Radical surgical resection remains the main treatment modality in the management of sarcomas. Local recurrence is the primary cause of mortality after resection in these patients [5]. Low tumor grade and smaller tumor size are both well-established prognostic factors and are significantly associated with improved survival. Negative microscopic margin status (R0) is associated with improved survival [5]. The therapeutic aim of surgery in patients with STS is complete macroscopic resection, ideally with negative microscopic margins. This may oblige the decision to resect the surrounding tissue by the surgeon involved or adhere to the tumor.

However, because of the large size of these tumors and the intraoperative difficulty in accessing all involved margins, this goal is difficult to achieve, often resulting in the presence of positive microscopic margins. Tumors that are located in a strategic anatomical location, as presented here, are difficult to manage, and the surgeon needs to decide whether the surrounding vital structures are involved in the tumor or can easily and safely be dissected away from the tumor.

The pseudocapsule consisting of surrounding tissue to the sarcomatous tumor dictates occasionally unblock resection. In proximity to strategic and vital organs, clinical judgment is necessary and a more conservative approach should be considered.

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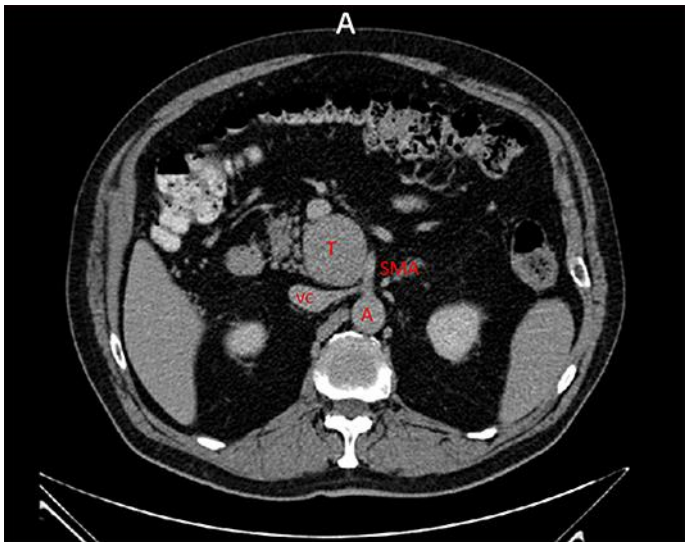

Fig. 1. The CT image depicts a soft density lesion adjacent to the root of the SMA and pancreatic head. T = Tumor; A =aorta; VC = vena cava.

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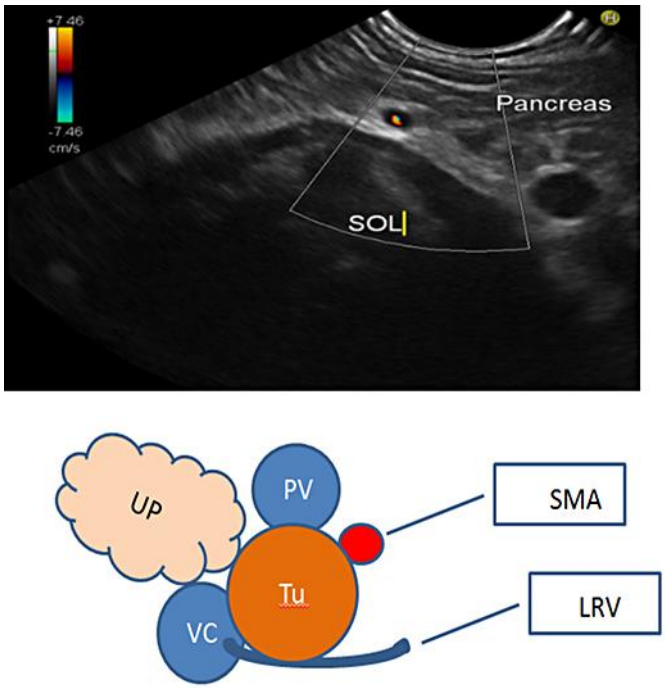

Fig. 2. **a** Sonographic image depicting the vicinity of the tumor to the uncinated process. **b** Diagram showing the strategic location of the tumor. SOL = Space occupying lesion; UP = uncinate process of the pancreas; PV = portal vein; VC = vena cava; Tu = tumor; LRV = left renal vein.